CASE REPORTS

A-V DISSOCIATION IN DYSTROPHIA MYOTONICA

BY

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Disturbance of cardiac rhythm in association with dystrophia myotonica has been recognized since Griffith (1911) recorded his finding of bradycardia in a patient with this disease. Since then other cases have been described and the subject has been reviewed by Spillane (1951) and Fisch (1951), the latter author concluding that of 85 cases of dystrophia myotonica 68 per cent had abnormal electrocardiograms and that of these 91 per cent were conduction defects and/or disturbances of rhythm.

The degree of cardiac involvement is generally slight but the case here reported was probably unique in presenting with Stokes-Adams attacks and terminating fatally.

Clinical Description

In 1941 Mrs. A. W., then aged 42 years, had an attack of unconsciousness of short duration. During the ensuing nine years further attacks of varying severity occurred at irregular intervals. There were no further symptoms of cardiovascular or other disease and no previous history of rheumatism or serious illness. The patient had never been pregnant.

An electrocardiogram in 1950 showed latent heart block (P-R., 0.28 sec.), and left bundle branch block. A diagnosis of Stokes-Adams attacks was made and treatment with atropin sulphate 1/100 of a grain by mouth four times daily was started. There was freedom from attacks for six weeks followed by their gradual return until in August 1951, their incidence was more frequent than ever and quite uninfluenced by atropin. She was admitted to hospital in February, 1952, and on the same day a typical Stokes-Adams attack was witnessed.

Examination of the cardiovascular system showed no signs of congestive failure. Peripheral arteries were clinically normal, pulse regular, rate 42, blood pressure 140/65. The heart was not enlarged, the cardiac impulse normal; the first heart sound was occasionally augmented, presumably by the auricular sound. The second heart sound was widely split in all areas. No murmurs were audible.

The facies was myopathic and bilateral ptosis was present; the speech was slurred and monotonous. There was muscular wasting, severe in the sterno-mastoid, spinati, and trapezii, and moderate in the muscles of the upper arms and thighs and associated with appropriate weakness. The grip was myotonic, the tendon reflexes diminished generally, and the thyroid gland diffusely enlarged.

Serum Wassermann and Kahn reactions were negative and lumbar puncture normal; slight left ventricular enlargement was demonstrated by teloradiogram. Gastrocnemius muscle biopsy showed considerable interstitial accumulation of adipose tissue cells which separated the bundles of muscle fibres. The latter showed considerable variation in size, and many were atrophic; a very small proportion of hypertrophied fibres were present. In a few muscle fibres were rows of centrally placed nuclei. Neither striated annulets nor excess of sarcoplasm was seen in relation to any muscle fibre. The appearances corresponded with those described by Wohlfart (1951), although not all the features that he mentioned were present.

Electrocardiograms showed varying degrees of A-V conduction from sinus bradycardia (Fig. 1 A), to complete A-V dissociation (Fig. 1B). The shape of the ventricular complexes varied, being at all times abnormal and changing indiscriminately from the appearances of left bundle branch block to those of right bundle branch block (Fig. 1C). Stokes-Adams attacks continued to occur frequently while the patient was in hospital and a cardiogram made during one such episode showed probable ventricular tachycardia which, on recovery, was followed by complete A-V dissociation (Fig. 1D).

In an attempt to control the incidence of ventricular tachycardia it was decided that a course of quinidine sulphate should be given. The test dose of 3 grains was followed by such severe tinnitus and increased

frequency of syncopal attacks that use of the drug was abandoned. On March 20, treatment with ephedrine hydrochloride, $\frac{1}{4}$ of a grain t.i.d. was started and immediately the syncopal attacks stopped. At the end of April they reappeared and in one of them at home the patient died. We have ascertained that she had stopped taking ephedrine. No post-mortem examination of the body was made.

FIG. 1.—(A) Sinus bradycardia at 36 a minute. P-R, 0.26 sec., and left bundle branch block. (B) Complete A-V dissociation, auricular rate 133, ventricular rate 46; auricular rhythm not quite regular. (C) Complete A-V dissociation; ventricular complexes of varying type. (D) Electrocardiogram taken toward the end of a Stokes-Adams attack showing ventricular tachycardia, rate 170, followed by complete A-V dissociation.

Discussion

Although the main impact of dystrophia myotonica is usually upon skeletal muscles, in the case described the symptoms pointed to early severe heart involvement; only much later were there any complaints referable to the neuro-muscular system. It might be argued that the patient's preoccupation with the syncopal attacks explain why the comparatively minor disabilities of motor weakness and myotonia were overlooked. On the other hand, the disease may exist in the absence of symptoms, as was demonstrated by the patient's brother who, although declaring himself to be well and strong, had signs of advanced dystrophia myotonica.

Reports of such severe cardiac damage as was seen in this case are few. Brain (1951) mentions the occurrence of complete heart block and one case from a series reported by Evans (1944) had two probable Stokes-Adams attacks, a long P-R interval (0.30 sec.), and atypical bundle branch block. Broustet (1945) describes auricular flutter with A-V dissociation in the presence of an atypical myopathy; the significance of the association in this case is doubtful, however, as the patient had hypertension.

Although in our case disturbance of A-V conduction is undeniable, caution must be exercised in interpreting the abnormal QRS complexes as evidence of one or other type of bundle branch block. It is more likely that the variation in complexes indicated the establishment of ectopic foci of stimulation in one or other ventricle. The labile nature of the cardiographic changes in this case may, perhaps, indicate that errors of cardiac conduction in dystrophia myotonica represent functional change rather than severe myocardial disorganization.

Summary

A case of dystrophia myotonica in which Stokes-Adams attacks were early manifestations of the condition is described. The attacks were due to the onset of abnormal ventricular rhythm in the course of A-V dissociation. The absence of other causes of cardiac disease enhances the probability that the heart block was related to the dystrophy, the heart lesion being of unusual severity.

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